A43.E413
JACC March 9, 2010
Volume 55, issue 10A

CONGENITAL CARDIOLOGY SOLUTIONS
(PEDIATRIC CARDIOLOGY AND ADULT CONGENITAL HEART DISEASE)

TOTAL ANOMALOUS PULMONARY VENOUS RETURN--IMPACT OF PREOPERATIVE OBSTRUCTION AND EARLY ELECTIVE REPAIR ON OUTCOME AND NEED FOR PROLONGED MECHANICAL VENTILATORY SUPPORT

ACC Poster Contributions
Georgia World Congress Center, Hall B5
Monday, March 15, 2010, 9:30 a.m.-10:30 a.m.

Session Title: Pediatric Cardiology
Abstract Category: Pediatric Cardiology
Presentation Number: 1174-403

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Background: Total anomalous pulmonary venous return (TAPVR) requires surgical repair during infancy, but timing of elective repair when the anomalous pathway is unobstructed has not been clarified.

Methods: Retrospective review of all infants diagnosed with TAPVR as an isolated lesion at Children's Hospital of Wisconsin from 1991-2007 was performed to assess location of drainage, presence of obstruction, age at presentation, age at surgery, death, need for ECMO, length of hospital stay (LOS), length of mechanical ventilation (MV), and late pulmonary venous obstruction.

Results: 65 pts were identified, with 38 (59%) draining supracardiac (SC), 10 (15%) cardiac, 11 (17%) infracardiac (IC), and 6 (9%) mixed (M). 50% presented in the 1st 24 hrs of life with a range of 0-188 days. There were 3 early (4.6%) and 5 late deaths (7.7%) after surgery, all in pts with obstruction preop (5 SC, 3 IC); 4/5 late deaths occurred in pts who developed recurrent obstruction. 39/65 had obstruction identified preop (60%); 25 SC,11 IC, and 3 M, with median LOS and MV of 18 and 8 days. 8/39 needed ECMO support after surgery; 4/8 survived. 26/65 (40%) had no obstruction preop and none died, required ECMO support after surgery, or developed late obstruction with a median LOS and MV of 13 and 7 days. Of the 26 pts without obstruction, timing of surgery was elective at the discretion of the supervising cardiologist with 15/26 having surgery <10 days after presentation (median age 18d); 53% (8/15) of these pts had MV <5 days. In contrast, of the 11 pts who had elective surgery >10 days after presentation (median age of 56 d), 100% required MV for >5days (p=0.007 by Chi-Square analysis).

Conclusions: Isolated TAPVR appears to be at highest risk for death, need for ECMO support, and late postop obstruction when obstruction is present preop. Pts with unobstructive TAPVR do very well, but potential morbidity related to prolonged MV appears to be significantly reduced by early elective surgery. We speculate that delayed elective surgery in this group extends abnormal LA/LV filling prior to repair and results in altered chamber compliance that necessitates prolonged MV as the left heart remodels after surgery.